Case Report
JCAM

A Rare Clinical Condition: Erasmus Syndrome

Nadir Görülen Klinik Bir Durum; Erasmus Sendromu

Erasmus Syndrome

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Özet

Sistemik Skleroz (SS), cildin ve iç organların fibrozisi ile giden, sebebi tam olarak aydınlatılamayan sistemik otoimmün bir hastalıktır. Hastalığın çevresel faktörlerle ilişkisi olduğu bilinmektedir. Özellikle silica tozlarıyla maruziyetin bir takım immun reaksiyonların tetiklenmesiyle hastalığın patogenezinde rol aldığı düşünülmektedir. Silikozis ve SS birlikteliği Erasmus Sendromu (ES) olarak tanımlanmaktadır. Burada 6 yıldır silikozis nedeniyle takipli, kotlama işçisi olarak çalışan 30 yaşında sistemik sklerozlu bir olgu sunulmaktadır.

Anahtar Kelimeler

Silikozis; Sistemik Skleroz; Erasmus Sendromu

Abstract

Systemic sclerosis (SS) is a systemic autoimmune disease progressing with fibrosis of the skin and internal organs, the cause of which cannot be precisely explained. The disease is known to be associated with environmental factors. In particular, exposure to silica powders is believed to have a part in the pathogenesis of the disease by the triggering of a number of immune reactions. Silicosis and SS association is defined as Erasmus Syndrome (ES). Here, we report on a 30-year-old patient working in denim sandblasting who developed SS while being followed for 6 years due to silicosis.

Keywords

Silicosis; Systemic Sclerosis; Erasmus Syndrome

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Introduction

Systemic sclerosis (SS) is not only a disease progressing with skin involvement but also a systemic autoimmune disease which affects internal organs such as the gastrointestinal system, kidney, lung, and heart, and which is characterized by fibrosis [1]. The etiopathogenesis of the disease is not yet fully understood. However, vinyl chloride, organic solvents, benzene, and silica powders have been shown to be associated with the disease [2]. Silicosis is an incurable disease characterized by an irreversible and progressive fibrotic reaction in the lung tissue due to silica crystals of respirable size. Silica is a mineral that occurs intensively in the earth's crust. Exposure to silica powders is high in people workingin mines and quarries, ceramic manufacturing, pottery making, and denim sandblasting works [3]. The association between silicosis and SS is defined as Erasmus Syndrome (ES). Here, we report on a 30-year-old patient working in denim sandblasting who developed SS while being followed for 6 years due to silicosis.

Case Report

The patient, who was working in denim sandblasting for 10 years and had been followed with a diagnosis of silicosis for 6 years, applied to the rheumatology clinic with whitening and bruising on the hands in response to cold exposure and pain in the hand joints. The patient started to have pains that increased especially at nights, accompanied by swellings in the wrists and metacarpophalangeal joints. During the previous 3 weeks, a necrotic skin ulcer had occurred on the second finger of the left hand of the patient, who described recent hardening of the skin. The patient had no relevant features in the family history. Physical examination found TA of 130/80 mmHg and body temperature of 36.7°C. There were telangiectasias on the face and vertical lines on the lips. The mouth opening was narrowed. Bilateral fine crackles were found in breathing sounds. While hardening of the skin was detected on the back of the hand, a digital ulcer was detected on the second finger of the left hand (Figure 1). In the thoracic tomography, while enlarged multiple lymph nodes measuring more than 40 mm in maximum size were observed in the mediastinum and subcarinal space, septal thickening was observed in both lung bases (Figure 2). In the transthoracic echocardiography, pulmonary artery pressure was detected as 26 mmHg and EF was detected as 65%. In pulmonary function test, FEV1, FVC, FEV1/FVC, and DLCO were 65%, 72%, 71%, and 79%, respectively. The sputum examination and culture for acid resistant bacillus (ARB) were negative and a tuberculin skin test (PPD) was measured as 7 mm. In the laboratory examination the following values were detected: WBC: 8400/mm3, platelet: 365 × 103 per µL, Hb: 12.2 gr/dl, MCV: 81, C-reactive protein: 78 mg/dl (N:0-3), erythrocyte sedimentation rate: 45 mm/h. Immunological investigations showed rheumatoid factor (RF): 34 IU/l (N:0-15), anti cyclic citrullinated peptide (anti-CCP): 5 (N:0-7), antinuclear antibody (ANA) homogeneous and granular pattern 3+ with anti SCL 70. The patient was diagnosed with SS based on the available clinical and laboratory results. The disease was considered as ES due to the progression of the silicosis. Lung involvement of SS was scanned by high resolution computed tomography but neither honeycomb nor ground glass opacity were detected. While methotrexate

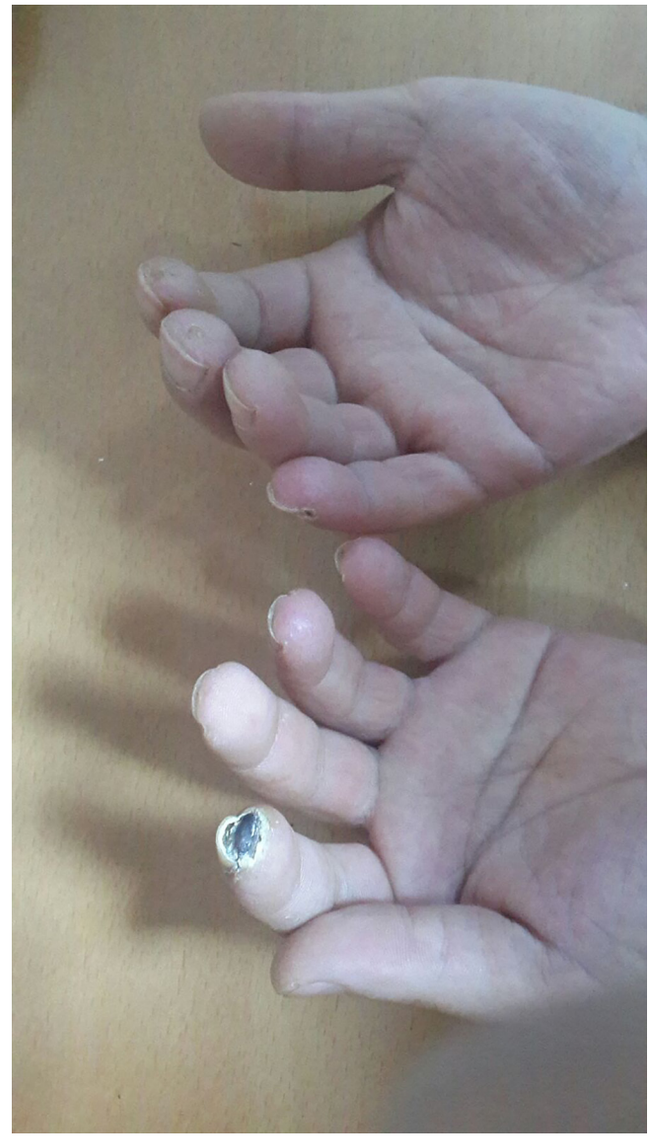

Figure 1. Digital ulcer on the 2nd finger of the left hand

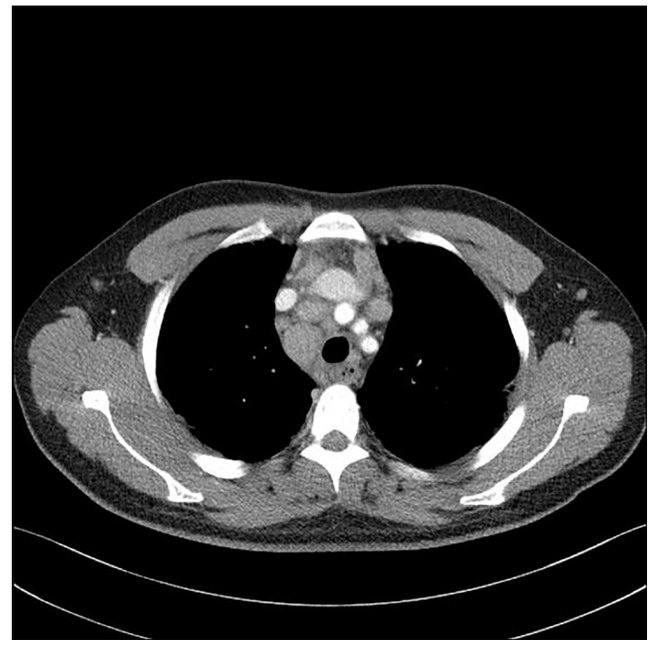

Figure 2.Thoracic tomography; multiple lymph nodes in the mediastinum

15 mg/week was administered for the systemic disease of the patient, calcium channel blocker, pentoxifylline, acetylsalicylic

acid, and intravenous ilioprost treatment were administered for the digital ulcer. Recovering from the digital ulcer during follow-ups, the patient was discharged with recommendations.

Discussion

Erasmus Syndrome was first reported by Erasmus in 1957 and its relationship between silicosis and SS was revealed. In addition, silicosis was also reported to be associated with pulmonary tuberculosis, lung cancer, rheumatoid arthritis, and systemic lupus erythematosus [4]. Although the pathogenesis of the relationship between silica powders and SS development has not been precisely explained yet, it is thought that silica particles are phagocytosed by macrophages, which ultimately leads to the release of lymphokine and chemokine by activating the fibroblasts. This also increases the synthesis of collagen and glycosaminoglycan and suppresses cellular immunity [5]. Also, Otsuki et al. reported that fasand caspase 8 antibodies and the lymphocyte-mediated apoptosis play a key role in tissue damage and immunity [6].

In SS cases developing secondary to silicosis, autoantibodies such as anti-scl 70 and RF are generally detected as positive; however, no clinical difference from the classical SS is observed [5]. Also in our patient, anti-scl 70 and RF were positive. The association of silica powders with tuberculosis has been known for many years. The incidence of tuberculosis in patients with silicosis is higher than in people without silicosis. The necessary examinations should be carried out when this diagnosis is suspected, because there is not always clear clinical evidence. In our patient, PPD was performed to rule out tuberculosis, and ARB and sputum culture were performed. However, there were no positive findings.

In SS cases developing with silicosis, decrease in diffusion capacity in the lung is remarkable. Respiratory insufficiency is generally seen in the form of restrictive pattern; dry cough and exertional dyspnea constitute the clinical spectrum [7]. Our patient's pulmonary function test was in restrictive pattern and there was a slight decrease in diffusion capacity. While the risk of secondary malignancy development increased with connective tissue disease, the risk of lung canceris further increased in patients with SS when in association with silica powders [8]. Therefore, attention should be paid to secondary malignancy in patients with ES and the necessary periodic examinations should be performed.

In silicosis-related SS cases, treatment does not vary much; it includes immunosuppressives such as corticosteroids, cyclophosphamide, and azathioprine [3]. In patients with secondary complications of SS such as Raynaud's phenomenon, digital ulcer, gastrointestinal and cardiovascular system involvement, palliative treatments for these involvements are also used. Thus, because digital ulcer was at the forefront in our patient, treatment for this involvement was given.

Because silicosis is a refractory disease that occurs as a result of exposure to silica, connective tissue diseases may develop in these patients. Especially, attention should be paid to the possibility of rarely-seen ES and periodic follow-ups should be performed with respect to potential lung cancer.

Competing interests

The authors declare that they have no competing interests.

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